197 Clinical follow up after liver transplantation in children with cystic fibrosis

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Background: 2–10% of children with cystic fibrosis (CF) develop hepatic cirrhosis with portal hypertension. The liver disease is a significant cause of morbidity and mortality and isolated liver transplantation (LTX) is an excepted therapeutic option. The effect of LTX on clinical parameters like pulmonary function is still under discussion.
Patients and Methods: We evaluated clinical parameters of 9 children with CF, at an age (mean±SD) of 14.1±3 years, before and one year after LTX.
Results: Before liver transplantation all patients had a mild to moderate pulmonary disease: FVC 92.2±16% (71.4–119%), FEV1 88.7±16.1% (68–103%), MEF25 61.7± 28.5% (35–114%), CN Score 8±4.3 points. They showed a malnutrition: BMI 17.2±1.8% (15.3%-20.3%), upper arm circumference 17.8±0.9 cm, triceps skin fat fold 5.2±1.2 cm, 13.5±7.2% of bodyweight were fat. One year after LTX all patients were in stable pulmonary situation: FVC 93.9±14% (75.5–114%), FEV1 88.6±12.9% (68–108%), MEF25 61.6±30.4% (24.7–106%), CN Score 7.5±4.8 points. There weren't any significant changes in the airway microbiology under immunosuppressive therapy (steroid, Ciclosporin±Basilixumab), the serum IgG levels declined significant from 17.4±3.9 g/l to 9.8±2.3/l. The growth over the year was 6.9±5 cm and the increase of weight come to 5.1±6.1 kg, while the BMI didn't increase in this first year after transplantation. Certainly we found an increase of the body fat mass (to 18.2±3.8% of body weight), of the upper arm circumference (to 20±4.2 cm) und of the Triceps skin fat fold (to 11.4± 8.1 cm).
Conclusions: Liver transplantation is an effective therapy in children with CF related liver disease and can stabilise pulmonary function and improve nutritional status in patients with CF and mild or moderate pulmonary involvement before LTX.

198 Consideration on ursodeoxicholic acid treatment

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Aim study was to comparatively assess the value of the ursodeoxycholic acid (UDCA) treatment in cystic fibrosis associated liver disease (CFLD).
Methods: Study lot included 26 children diagnosed with CFLD-27.95% from all 93 children followed up in our centre. Comparative analysis was done between 2 study groups: group 1 (14 patients, median age at diagnosis 6.6 years), treated with UDCA; group 2 (12 patients, median age at diagnosis = 14.8 yrs), who did not receive UDCA. Data were retrospectively evaluated for a period of 2 years (2004–2006). Groups were aged matched. We assessed the following data: clinical status, liver function tests (LFT), cholestasis-linked enzymes, abdominal ultrasound (US) exam, and, in some cases: hepatobiliary scintigraphy, cholangioMRI, liver biopsy. Patients from group 1 were treated with 20 mg/kilo body weight UDCA.
Results: Among group1, 78.57% (n = 11) patients responded to treatment by normalization of the LTF and cholestasis-linked enzymes. Better clinical status was also certified accompanied by improvement of imagistic data. Only 21.42% from patients treated with UDCA developed severe LD, comparative to 58.33% untreated patients. Among group 2, the delay of the diagnosis had a large negative impact on disease's natural history. All 12 patients presented with at least one complication; 7 patients with portal hypertension. We have noted worsening of clinical condition and deterioration of biochemistry and liver imaging in the untreated patients.
Conclusion: The better evolution of the patients treated with UDCA sustain its recommendation, from the very early diagnosis of liver involvment.

199 Gallbladder anomalies among children with CF

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Aim study is to evaluate the prevalence of gallbladder anomalies in CF children and its correlation with cystic fibrosis associated liver disease (CFLD).
Methods: Ninety-three hildren aged between 3 month-18 years were evaluated by abdominal ultrasound examination (US), hepatobiliary scintigraphy and MRI cholangiography for detection of gallbladder anomalies. Cholestasis-indicating enzymes, bilirubinemia and liver function test (LFT) were recorded as well. Clinical examination with a comprehensive anamnesis was performed every visit. Pursued gallbladder anomalies: microgallbladder, cholelithiasis and cholecistitis.
Results: Gallbladder's anomalies were detected in 36 cases (38.7%). Microgallbladder was the most frequent manifestation – 34.4% (32 patients). Cholelithiasis occurred in 12 patients (12.9%); in 10 cases microgallbladder was associated (83.3% reported to cholelitiasis cases). Acalculous cholecystitis was documented in only 2 patients (2.1%). Liver disease was diagnosed in 26 patients (27.95%); five of them (19.23%) associated gallbladder anomalies. Among 36 patients with gallbladder anomalies only 13.88% had liver disease.
Conclusions: Gallbladder disease is a common problem in CF children, with an increasing incidence. The most prevalent morphological change is microgallbladder. Cholelithiasis occurs frequently in children with microgallbladder. We have noticed, without finding a consistent explanation in this issue, that the gallbladder anomalies are rare in patients with CFLD and liver disease is uncommon in children with gallbladder anomalies.